Monochorionic-diamniotic twin pregnancy complicated by twin reversed arterial perfusion sequence and retroplacental hematoma – a case report

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Abstract
Twin reversed arterial perfusion (TRAP) sequence is a rare and severe complication specific to monochorionic twin pregnancies, involving the presence of an acardiac twin and a structurally normal co-twin (pump twin). We report on the case of a 33-year-old female with a biamniotic monochorionic twin pregnancy complicated with TRAP sequence and polyhydramnios. The patient underwent fetoscopic termination of the acardiac twin and at 34 gestational weeks (GW) was readmitted with a retroplacental hematoma. The patient gave birth through caesarean section to a living female fetus, weighing 1480 g. To the best of our knowledge, this is the first case reporting a twin pregnancy with TRAP sequence complicated with retroplacental hematoma.

Keywords: twin pregnancy; acardiac twin; TRAP sequence

Monochorionic twin pregnancies represent a challenge for the obstetrician, due to the high risk of vascular placental anastomoses development, leading to communication between the two fetoplacental systems. This abnormal communication is often responsible for an imbalanced blood supply between the two fetuses, resulting in the twin-to-twin transfusion syndrome (TTTS) [1]. The extreme form of inter-twin vascular anastomoses is the twin reversed arterial perfusion (TRAP) sequence, a rare (1/35,000 pregnancies) and severe complication specific to monochorionic multiple pregnancies [2]. TRAP sequence involves the presence of a acardiac twin and a structurally normal co-twin (pump twin). The normal twin has a high mortality rate (50-70%), especially due to congestive heart failure [1]. The purpose of this paper is to present the ultrasound findings in a twin pregnancy complicated by TRAP sequence and retroplacental hematoma.

Case report
We report the case of a 33-year-old female referred to our institution at 24 GW with the initial diagnosis of a monochorionic twin pregnancy with one normally developed alive fetus, and one dead fetus. Ultrasound revealed 24GW intrauterine monochorionic-diamniotic (MoDi) twin pregnancy with a structurally normal twin, accompanied by a mass presenting an amorphous abdominal cavity, two abnormal lower limbs and no sign of head, upper limbs, thorax or cardiac structure (fig 1a-c).

Polyhydramnios was present, with the largest amniotic pocket measuring 141.8 mm (fig 1d). Initial Doppler evaluation of the normal twin indicated no signs of cardiac damage. Doppler imaging showed pathognomonic reversed arterial perfusion from the pump twin to the acardiac twin, through a single umbilical artery. We established the diagnosis of 24 GW MoDi twin pregnancy with TRAP sequence and polyhydramnios, and we performed amniocentesis for both diagnostic purpose...
(revealing normal karyotype) and therapeutic intention. Fetoscopy was performed, using laser photoocoagulation of the umbilical cord of the acardiac twin, followed by decompression amniocentesis.

The patient was readmitted to our institution at 34 GW for preterm premature rupture of membranes (PPROM). Ultrasound revealed intrauterine growth restriction (IUGR) of the living twin, corresponding to 30GW, the acardiac fetus present at the inferior pole of the uterus, with arrested development, and a marginal retroplacental hematoma (RH), measuring 67/24 mm, with no Doppler signal. Five hours later, the patient was reassessed by ultrasonography, showing a decrease in the amniotic fluid volume, as compared to the previous examination, and an increase in the size of the RH, reaching 87.5/32.8 mm. Fetal extraction was immediately decided, and the patient gave birth through caesarean section to a living female fetus, weighing 1480 g, Apgar score 6 at 1 minute.

Pathologic examination revealed an acardius acephalus fetus with the absence of the entire upper extremity, abdomen and inferior limbs with normal appearance, external genitalia with ambiguous differentiation, absence of anal orifice. The umbilical cord of the acardiac twin presented two vessels, a cystic structure and thrombosis, following laser photoocoagulation. Anastomoses were present between the two umbilical cords.

Discussions

TRAP sequence is the extreme form of TTTS, with the underlying pathophysiological mechanisms still incompletely understood. In a case with TRAP sequence, there is a normal twin which becomes the pump, its heart supporting not only its own blood flow, but also the acardiac twins’ one [3]. The pump twin will eventually develop arterial-steal phenomenon, characterized by progressive cardiac insufficiency, polyhydramnios and consequently heart failure [4]. Diagnostic amniocentesis is mandatory, in order to rule out chromosomal anomalies in the pump twin [5].

Particular complications encountered in our case, that have not been specifically described in the literature so far, to the best of our knowledge, were IUGR of the pump twin (34 corresponding to 31 gestational weeks) and RH. In the largest series on the outcome of 49 acardiac twin pregnancies without intrauterine surgical treatment the mortality rate was 51% and only 24% of the patients delivered after 36 gestational weeks [6]. Other studies described a 50-70% mortality rate without adequate treatment [7,8].

Misdiagnosing TRAP sequence for twin pregnancy with intrauterine fetal demise of one twin is a rather common mistake with less experienced obstetric sonographers, reported by Weisz et al [9] in all 6 cases of acardiac twins.

Fig 1. a) fetal face and b) four chambers view – normally developed heart in the structurally normal twin; c) vascularization of the acardiac twin; d) polyhydramnios; e) measurement of the femur length of the pump twin, corresponding to 30w + 2d, revealing intrauterine growth restriction; f) measurement of the femur length of the acardiac twin, corresponding to 21w, demonstrating the arrested development as a consequence of the laser photoocoagulation of the umbilical cord.
that they described. TRAP sequence should be suspected in MoDi pregnancies when one fetus is morphologically normal, whereas the other lacks cardiac structures and/or activity [10]. The typical ultrasonographic appearance of the acardiac twin is a hydropic mass without cardiac activity, which continues to grow [6]. Arterial flow in the umbilical artery of the acardiac twin makes the diagnostic certain [2]. Doppler imaging shows pathognomonic reversed arterial perfusion from the pump twin to the acardiac twin. These features may be detected as early as first trimester of pregnancy [6]. In our case, pathognomonic Doppler findings – reversed arterial perfusion, arterial flow in a single umbilical artery to the acardiac twin – were noticed at our initial evaluation, which was performed in the second trimester. Ultrasound has also a major role of assessing poor prognostic factors, such as a rapidly growing hydropic and large acardiac twin, the weight ratio acardiac/pump twin higher than 0.7, polyhydramnios and signs of cardiac failure of the pump twin (reversed diastolic flow in the umbilical artery, umbilical vein’s pulsatile flow, reversed flow on the ductus venosus) [2,6].

In case of ultrasound alarm signs or prophylactically, as preferred by some medical centers [11], interventional techniques can be used. Laser coagulation of the umbilical cord was the treatment of choice in our case, since ultrasound high-risk signs were noted in the course of pregnancy. The interruption of the blood flow to the acardiac twin allowed the normal twin to further develop, suppressing the cardiac stress and preventing further hemodynamic disorders.

In conclusion, TRAP sequence is a rare entity, requiring early diagnosis and careful ultrasound monitoring, in order to select the best management option for the pump twin. To the best of our knowledge, this is the first case reporting a twin pregnancy with TRAP sequence complicated with RH.

References